Turkish Journal of Cerebrovascular Diseases 2024; 30(1): 51-53 Turk J Cereb Vasc Dis doi: 10.5505/tbdhd.2023.37132

LETTER TO EDITOR

EDİTÖRE MEKTUP

UNILATERAL LEUKOENCEPHALOPATHY IN PARRY ROMBERG SYNDROME: HIGH-FIELD MAGNETIC

RESONANCE IMAGING FEATURES

PARRY ROMBERG SENDROMUNDA TEK TARAFLI LÖKOENSEFALOPATI: YÜKSEK ALAN MANYETIK

REZONANS GÖRÜNTÜLEME ÖZELLİKLERİ

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Keywords: Ischemic leukoencephalopathy, non-inflammatory arteriopathy, perfusion, sympathetic. **Anahtar Sözcükler:** İskemik lökoensefalopati, inflamatuar olmayan arteriyopati, perfüzyon, sempatik.

Dear editor,

Multimodal magnetic resonance imaging was performed on a 41-year-old male patient due to atypical migrainous headaches and short-duration episodes of bilateral dysesthetic attacks that had been ongoing for at least ten years. MRI revealed highly diagnostic features of Parry-Romberg syndrome (Figure).

The patient's physical examination revealed mild but significant right facial hemiatrophy, which has not previously been specifically complained about, noticed or categorized. Sleep and wakefulness EEG were normal. The patient, who also complained of arthralgia, was diagnosed as an undifferentiated connective tissue disease because the rheumatological tests performed in this regard were not guiding.

Parry Romberg syndrome is a very rare but typical syndrome, and being familiar with brain MRI findings can prevent unnecessary tests on the patient. Our aim is to remind the readers of the Journal about the syndrome and its MRI features with this case vignette.

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Please cite this article as following: Çakan M, Topçuoğlu MA, Göçmen R, Unilateral leukoencephalopathy in Parry Romberg Syndrome: High-field magnetic resonance imaging features. Turkish Journal of Cerebrovascular Diseases 2024; 30(1): 51-53. doi: <u>10.5505/tbdhd.2023.37132</u>

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Figure. A: Coronal T2-weighted MR imaging sequence: It has been documented that the right frontoparietal inter-diploic distance is asymmetrically narrowed (Red arrows), causing dysmorphism in the form of an easily noticeable reduction in the head and forehead size in the vertex (White arrows). Diffuse and confluent white matter hyperintensity is observed in the centrum semiovale. Additionally, it is accompanied by unilateral sulcal effacement. This phenomenon has been described in Parry Romberg syndrome patients due to cerebrocortical blood volume increase from recent seizures or migraines(1,2). Sulcal effacement is seen in approximately 6% of the syndrome (1). Technical Details: Repetition Time (TR): 4923 msec; Echo Time (TE): 100.408 msec; Magnetic Field Strength: 3 A/m; Slice Thickness (ST): 4 mm.

B. Axial FLAIR (Fluid-attenuated inversion recovery) sequence: The confluent hyperintense lesion in the subcortical white matter is seen in the horizontal plane and is limited exclusively to the right side. This type of strictly unilateral location of the white matter lesion is seen in one-third of the cases(1). Sulcal effacement; decreased horizontal diameter of the intracranial dome; and a relatively narrowed diploe distance are all visible on the right side. Technical details: Repetition Time (TR): 10,000 msec; Echo Time (TE): 88.128 msec; Magnetic Field Strength: 3 A/m; Slice Thickness (ST): 4 mm.

C. Postcontrast T1-weighted MR imaging reveals leptomeningeal (open red arrows) perivascular enhancement and sulcal effacement. The T2 hyperintense lesion appears T1 hypointense and has a heterogeneous texture. This type of development is rare but well described in Perry Romberg syndrome. The cause has been assumed to be cerebral progressive proliferative arteriopathy or autoimmune neurovasculitis, but this has not been fully confirmed. It has been claimed that inflammation is present at least during some period of the disease course. Later, leptomeningeal and cortical blood vessel endothelial degeneration, hyalinizing changes, and partial obliteration develop. Possibly endothelial regeneration is not complete. (3,4). Technical details: Repetition Time (TR): 8684 msec; Echo Time (TE): 3272 msec; Magnetic Field Strength: 3 A/m; Slice Thickness (ST): 1 mm.

D. Head computerized tomography (CT) in bone density: Frontal and parietal bone atrophy are documented (White arrows).

E-F-G: MR Perfusion: Cerebral blood flow (CBF, E), Cerebral blood volume (CBV, F) and time-to-peak mapping (TTP, G) are compatible with hypoperfusion (CBF and CBV decreased, TTP prolonged) in the lesioned white matter area, while preserved or relatively increased perfusion in the cortex is notable. This finding has been reported in previous SPECT studies (3).

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Unilateral leukoencephalopathy in Parry Romberg Syndrome

Ethics

Ethics Committee Approval: Since this paper is a letter to the editor, ethics committee approval is not required.

Informed Consent: The authors declared that informed

consent form was signed by the patient. Copyright Transfer Form: Copyright Transfer Form was signed by the authors.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices: MÇ, MAT, RG. Concept: MÇ, MAT, RG. Design: MÇ, MAT, RG. Data Collection or Processing: MÇ, MAT, RG. Analysis or Interpretation: MÇ, MAT, RG. Literature Search: MÇ, MAT, RG. Writing: MÇ, MAT, RG.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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